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RESEARCH PAPER

Stumbling, struggling, and shame due to spasticity: a qualitative study of adult persons with hereditary spastic paraplegia

Hans C. J. W. Kerstens[a,b], Ton Satink[b], Maarten J. Nijkrake[c], Bert J. M. De Swart[b,c], Bas J. H. Van Lith[c], Alexander C. H. Geurts[c] and Maria W. G. Nijhuis-van der Sanden[a,c]

[a]Radboud University Medical Center, Radboud Institute for Health Sciences, IQ Healthcare, Nijmegen, The Netherlands; [b]HAN University of Applied Sciences, Nijmegen, The Netherlands; [c]Radboud University Medical Center, Donders Institute for Brain, Cognition and Behaviour, Department of Rehabilitation, Nijmegen, The Netherlands

ABSTRACT

Purpose: Little is known concerning the impact of chronic spasticity on physical activities, social participation, and well-being, and whether patients' needs are addressed by current treatments. This study aims to investigate these lacunas in persons with a pure form of hereditary spastic paraplegia (HSP), in whom spasticity is a prominent symptom.

Methods: Fourteen patients with a pure form of HSP were interviewed. These interviews were recorded, verbally transcribed, and thematically analyzed.

Results: Four themes were identified which can be reflected by the phrases: (1) 'I stumble', (2) 'I struggle', (3) 'I feel ashamed', and (4) 'I need support'. Balance and gait problems led to limitations in domestic activities, employment, and recreation. 'Stumbling' also occurred due to pain, stiffness, and fatigue. Struggling was related to the continuous need for adaptation strategies, including the abandonment of some activities. Participants further reported feelings of shame, fear, and frustration. Lastly, they needed more support in daily activities than currently provided.

Conclusion: Besides treating spasticity-related motor impairments, patients with HSP need practical support for optimizing their physical activities and social participation. They also seek attention for the non-motor consequences of their chronic spasticity to improve their well-being. Patient-reported outcomes might help to address these needs.

➤ IMPLICATIONS FOR REHABILITATION

- According to patients with hereditary spastic paraplegia, interventions for spasticity should not only be aimed at reducing motor impairments, but also on reducing pain and fatigue, improving nighttime rest and general well-being, and optimizing the performance of relevant personal activities.
- Medical, role and emotional management in patients with hereditary spastic paraplegia can be improved only when individual needs are identified and monitored over the course of the disease.
- Besides assessment of bodily functions and physical capacities, systematic evaluation of patient-reported outcomes will help both patients and professionals to monitor the functional impact of disease progression and to evaluate the effects of interventions aimed at retarding this progression.

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Introduction

Dealing with the consequences of spasticity in daily life is challenging [1]. Current treatment focuses on reducing spasticity-related motor impairments and preventing secondary consequences [2]. A diversity of pharmacological, surgical, and physical interventions aim to reduce muscle tone, restore muscle balance across joints, and prevent secondary muscle changes, such as contractures [2–5]. Mainstay pharmacological treatment is either systemic (oral medication or intrathecal baclofen) or focal (e.g., intramuscular botulinum toxin injections). Besides pharmacological treatment, physical therapy may reduce the biomechanical consequences of spasticity, for example through muscle stretching exercises, (serial) casting, and learning to optimize the performance of activities [5].

The effects of the interventions mentioned above are commonly assessed with outcome measures at the physical impairments level [6], in particular with scales for muscle tone and measures of passive and/or active range of motion [7]. However, such outcomes do not assess the impact of spasticity on physical activities, social participation, or general well-being in persons with spasticity [3]. As a consequence, it is likely that the current medical treatment strategies, traditionally established by healthcare professionals, do not address all the needs and priorities of persons with chronic spasticity.

The primary aim of this study was to investigate how chronic spasticity impacts on physical activities, social participation, and well-being. The secondary aim was to identify whether patients' needs are adequately addressed by the current intervention

CONTACT Hans Kerstens Hans.kerstens@han.nl HAN University of Applied Sciences, Nijmegen 6525EN, The Netherlands

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strategies. In this study we focus on persons with a pure form of hereditary spastic paraplegia (HSP), who experience chronic, progressive spasticity of the lower limbs as their most prominent symptom. Leg muscle strength in these patients is relatively well preserved, probably because the reticulospinal descending tracts are able to compensate for the progressive axonal degeneration of the corticospinal tracts [8]. In addition, sensory deficits are usually mild and cognitive impairments absent [9]. Hence, investigating the impact of chronic spasticity in persons with pure HSP provides an opportunity to study the consequences experienced by and treatment needs in persons with 'relatively isolated' lower-limb spasticity.

Materials and methods

Design

A qualitative study was conducted between autumn 2015 and December 2017. A social constructivist epistemology was used in interactions with the interviewer to explore multiple perspectives of the realities perceived by patients with HSP [10]. Using this approach, the experiences of persons with HSP were interpreted with attention to the consequences of chronic lower-limb spasticity for physical activities, participation, and well-being, as well as to the needs of patients. The study was conducted using semi-structured interviews.

Participants

The participants were persons who had an established diagnosis of autosomal dominant HSP and who had a pure phenotype. Other inclusion criteria were being 18 years of age or older and having had spasticity for more than two years. One exclusion criterion was an inability to communicate in the Dutch language. For the inclusion of participants, purposive sampling was used [10]. This sampling method aims to capture the diversity of experiences and needs within the HSP population, which means that participants were selected to represent the target population regarding personal characteristics such as age, sex, severity of paraparesis and spasticity, and with regard to referral from different sites across the country. Participants were recruited from the database of the outpatient clinic of the department of Rehabilitation of the Radboud university medical center (Radboudumc) between 2014 and 2017. The Radboudumc is a nationally recognized center of expertise for HSP in the Netherlands. The rehabilitation physician at the Radboudumc contacted persons with HSP by telephone. If the person with HSP was willing to participate, information about the study was sent by email. If the person decided to participate, he or she was asked to contact the researcher by email. The primary researcher (HK) checked both the inclusion and exclusion criteria before an appointment for the interview at the participant's home was planned. All participants gave written informed consent prior to the interview. The local medical ethics committee approved the study protocol (registration number 2015–2121). All data were stored and reported in conformity with the Declaration of Helsinki [11].

Data collection

An interview guide was developed by the primary researcher (HK) and pre-tested among physiotherapy students for feasibility and length among physiotherapy students. The guide was not used as a strict checklist, but merely as a guideline (see Supplementary Material) to ensure completeness at the end of each interview [10]. The one-hour interviews started with open questions to gather information regarding spasticity and its experienced consequences. The in-depth questions focused on the impact of spasticity on physical activities, social participation, and well-being. Subsequently, the interviews focused on patients' reflections on the experienced benefits or adverse effects of the interventions they had received, and whether they felt that these interventions had been adequately tailored to their problems and needs. Next, the interviews explored patients' wishes and ideas about optimal spasticity management with a focus on the solutions they brought up themselves. The results of previous interviews were used to guide the interviews with subsequent participants. The interviews were digitally recorded, and during the interviews, key messages from the participants were written on sticky notes that were attached to a poster. These notes supported the conversations and enabled participants to verify the content obtained during the interviews. The poster with attached sticky notes was photographed to support the analysis after the interview. All interviews were conducted by the primary researcher (HK), a male physiotherapist with more than 20 years of clinical experience in the rehabilitation of patients with spasticity, including patients with HSP. All interviews were conducted at the participants' homes and two research assistants (physiotherapy students) provided support. These assistants registered the additional field notes and transcribed the audio recordings of interviews verbatim.

Data from the previous interviews was analyzed before the next interview was planned. The interviews were stopped at the point when new information no longer came up, in other words, when the collected information reached saturation [12]. To guaranty anonymity, personal details were removed from the interview transcripts and a unique numeric code was assigned to each interview before storing it. The key to identifying individual patients was stored in a separate data folder accessible only by HK.

Data analysis

The interviews were analyzed according to the principles of thematic analysis of Braun and Clarke (2006) [13]. The steps in this analysis comprise familiarizing oneself with the data, generating initial codes, searching for themes, reviewing themes, and defining themes [13]. The analysis followed an inductive approach without using any previously constructed framework for possible codes.

The principal investigator (HK) coded the first transcript and then discussed the coding process and initial codes with the subsequent coders (DV and TS), both experienced qualitative researchers. After consensus was reached, HK coded the second interview and, once again, discussed it with DV and TS. Again, after agreement was reached, HK coded the remaining interviews. TS peer-reviewed the procedure and the content of the analysis. The codes were compared, checked for overlap, and discussed until a consensus was reached.

During the next step, HK searched for themes by clustering the codes into potential themes. This clustering was supported by consulting the sticky notes attached to the posters. The clustering was discussed with TS until agreement was reached. After reviewing and defining themes, HK, TS and all other authors discussed the themes in three rounds until all approved the final themes. Data were analyzed by using Atlas.Ti version 8.1.27.0 (Scientific Software Development GmbH, Berlin, Germany).

Trustworthiness

We used the COREQ criteria for reporting qualitative research [14]. To ensure trustworthiness, the concept credibility (do the data and procedures address the intended focus, and do the themes cover the data?), dependability (do the data change over time?), and transferability (can the results be transferred to other settings or groups?) [15] were considered in a number of ways. Credibility was taken into account through purposive sampling. We aimed to sample a rich variety of experiences, and therefore included participants with different characteristics, for example, male and female, young and old, employed and unemployed. We presented the conversation notes to each of the participants so that they could check the accuracy. In addition, the principal investigator (HK) discussed the codes with DV and TS, and later in the process with all the researchers. Dependability was assessed using a pretested interview guide that was adapted according to the results of the previous interviews, to ensure that similar relevant topics were discussed in the subsequent interviews. To ensure transferability, the participant characteristics were described as well as the process of data collection and analysis.

Results

A saturation point for new information was reached after 11 interviews. To optimize credibility, we stopped including new participants after 14 interviews. In total, 15 patients with HSP were invited to participate, of whom one person refused due to personal circumstances. A caregiver was present during two of the interviews. The 14 participants comprised a heterogeneous group based on sex, age, marital status, employment, HSP subtype, disease duration, and on whether they had received regular physical therapy (see Table 1). Data analysis led to the identification of four themes that could be reflected by the phrases: (1) "I stumble", (2) "I struggle", (3) "I feel ashamed", and (4) "I need support". Each theme will be elaborated and supported by quotes from the participants.

"I Stumble"

All participants mentioned that they literally and/or metaphorically stumbled. We identified four categories that illustrated their stumbling: pain and stiffness, fatigue, impaired balance and gait, and day-to-day fluctuations. Each category is addressed below and introduced with a representative quote.

Sometimes the pain feels like I've been run over by a train (P2)

Almost all participants consistently mentioned leg pain as an important factor in their life. Some also experienced back pain, which they thought was caused by an altered posture and gait pattern. Pain often had an impact on the patients' emotions. One person expressed that sometimes she felt she could not live with the pain anymore: "I cannot explain it very well, but sometimes the pain is so unbearable that I would rather be dead" (P2).

Another complaint mentioned by all participants was leg stiffness. Both stiffness and pain in the legs increased the longer a sitting or lying position had to be maintained. Initiating walking after a seated activity (e.g., playing the accordion) or walking to the coffee machine during a work break was inconvenient due to the stiffness. Also, participants experienced leg pain and stiffness during the night that led to difficulties with their mobility in bed: "When I lie down, I am like a plank from my waist down. If I want to turn over, I have to move my legs with my hands and flip myself over" (P3).

I Am so tired that I could sleep standing up (P5)

Many participants expressed that living with spasticity was very taxing, leading to high levels of fatigue. They felt that, above all, their gait impairments required more effort and energy. Another presumed cause of fatigue was disturbed nighttime rest due to difficulties in finding a comfortable sleeping position as well as with rolling over in bed. To be able to perform their basic and work-related activities as well as possible, they went to bed early and took naps during the day. Furthermore, physical activity levels were set lower so that energy could be preserved and then devoted meticulously to the most important daily activities, for instance during valuable leisure time: "I do not enjoy walking, I only walk when it is really necessary. It exhausts me" (P6).

I Have difficulty with all the activities that require me to use my legs (P10)

All participants experienced problems with gait and balance control. They reported a gait pattern characterized by toe-walking and a diminished walking speed. They experienced a stumbling gait pattern and were afraid of falling or reported falls or near falls: "When I walk downhill, I need to tilt my torso backwards quite a bit to avoid falling" (P12). Due to both the impaired balance control and stumbling character of the gait, all standing and walking activities were considered by many as very energy demanding.

As a consequence, participants experienced limitations in a wide range of daily activities such as personal hygiene, housekeeping, gardening, employment, mobility including shopping

Table 1. Participant characteristics.

Participant	Sex	Age	Marital status	Employment	HSP Genotype	Disease duration	Regular physical therapy
1	Male	72	Single, children	No	SPG4	>20 years	Yes
2	Female	50	Single	No	SPG31	>20 years	No
3	Female	56	Cohabitant	No	SPG31	>20 years	No
4	Female	58	Married, children	Yes	SPG31	>20 years	Yes
5	Male	64	Married	No	AD-HSP	>20 years	Yes
6	Male	26	Single	Yes	SPG4	>10 years	Yes
7	Male	48	Married, children	No	SPG4	>10 years	No
8	Male	50	Single	No	SPG4	>10 years	Yes
9	Male	49	Cohabitant	Yes	SPG4	>20 years	No
10	Male	44	Cohabitant	Yes	AD-HSP	>35 years	Yes
11	Female	51	Married, children	Yes	SPG4	>6 years	No
12	Female	53	Married, children	Yes	SPG4	>18 years	No
13	Female	59	Married, children	Yes	SPG10	>4years	Yes
14	Female	66	Married	No	SPG4	>20years	Yes

and getting groceries, and leisure activities. They mentioned problems with standing still, regular walking, bending, and stepping across objects in addition to pain, stiffness, and fatigue as the factors underlying the limitations they experienced. They gave several examples to illustrate their problems in daily life. Participants explained that their personal hygiene was time-consuming and that it was difficult to complete the job in a standing position. Housekeeping was also problematic, in particular reaching for and cleaning low and high objects: "I need to step over the cable and the hose when I'm using the vacuum cleaner, and I cannot do that" (P3). Both housekeeping and gardening were fatiguing and demanded a lot of balance effort: "I used to trim the hedge myself. I can still do the trimming, but I can't manage to do the raking anymore. For that I need to be able to walk, and I can't anymore" (P1). Some participants reported that walking at work was difficult as well, because they could not concentrate on their gait. For example, it was hard to stand up regularly to get a printed document or a cup of coffee.

Regarding outdoor mobility, the activities of getting in and out of a car and using the gas or brake pedals were also reported by many as problematic: "I hope my driver's license will be renewed since using the brakes is getting problematic" (P8). Still, driving a car was very important to them to be able to visit people or go shopping. Only one participant did not have any problems driving a car. Riding a bicycle was also difficult because many participants were afraid of falling when using the brakes suddenly for stopping. Walking on an uneven surface, grit, or sand increased the risk of falling. Some people were accustomed to regular falls, but still feared that a fall one day might have serious physical consequences such as a bone fracture. Participants thought exercise and sports were important, but it was difficult to find a suitable sport given their physical limitations.

Some days I feel like a wreck, but sometimes I don't feel too bad at all (P2)

Although some participants related that their symptoms and activity limitations were relatively stable over a certain period of days or weeks, others reported strong and often unpredictable fluctuations from day to day. As one participant said: "When I walk to the car in the morning, I can feel whether it's going to be a good or a bad day. I don't know why that is, though" (P9). Fatigue and physical stress were mentioned by some participants as a possible explanation for these fluctuations, but others had no explanation: "Sometimes I walk 20 min through the village after work and have little discomfort, but I can't manage to do that again the day after" (P10). Besides day-to-day fluctuations, all participants experienced a gradual increase in disease progression, slowly leading to more activity limitations: "In the past, I could step over a gate, nowadays I have to be careful at a doorstep" (P2).

"I struggle"

All participants used adaptation strategies to compensate for their physical limitations, which made life a struggle to many. They either continuously adjusted the performance of an activity to their actual limitations over the years, or they ultimately decided to quit the activity completely. Each of these two categories is elaborated below and introduced with a representative quote.

I have to think before I do something: First I have to be standing steadily, then I can lift the groceries into the car (P13)

Participants used a wide variety of aids and physical home adaptations. They also asked for support from their spouses or other close kin during more strenuous activities such as housekeeping and gardening. Participants sometimes found practical solutions for everyday problems, for example, bending was avoided by picking up a metal object with a magnet or a shoe was picked up from the floor with a vacuum cleaner: "I use a vacuum cleaner to pick something up from the ground. I put the nozzle in a shoe so that I can lift it" (P7).

Participants regularly used walking aids such as a cane, walker, orthopedic shoes, or ankle-foot orthoses. Additionally, they used a trolley to walk better, safer, and to disguise their gait and balance difficulties: "I can walk well with a shopping trolley and no one notices I have a problem; I can imagine that I will need a walker later. But as long as I don't need to, I don't use one yet" (P11). For transportation across long distances, participants preferred cycling to walking and many used a power-assisted bike rather than a regular bike to save energy. Some, however, preferred riding a regular bike as long as possible to optimize and maintain their physical endurance level.

Another adaptation strategy was to increase the level of conscious attention to perform certain activities. Participants walked with increased visual control by watching the floor to be able to anticipate and avoid obstacles. Some explored unfamiliar target areas during the day or at quiet moments in anticipation of nighttime activities or crowded places: "During the day I check the surroundings so that I will know where the obstacles are when it is dark, or when it is very busy" (P7). While riding a bicycle, they searched for a quiet place to slow down and safely dismount from their bike. They planned their shopping route in advance to reduce the walking distance and to save as much energy as possible. Lifting heavy groceries into the car demanded preparation for stable standing to prevent falling.

Even though most adaptation strategies required increased attention and mental awareness, participants felt that, on the one hand, these strategies enabled them to keep performing important activities, which gave them a feeling of independence. On the other hand, all participants expressed that applying adaptation strategies had a huge impact on their daily lives: "I would love to not have to think about it when I go somewhere. For instance, what will we be doing on a staff outing? Will I be able to keep up, and will I be able to sit down somewhere along the way to rest? Can I park near my destination or do I still need to walk a certain distance?" (P4). In addition, finding the right balance between being active and taking rest was considered very difficult. While insufficient movement increased stiffness and hampered gait, too much physical activity resulted in reduced leg stability and loss of balance.

Diving is now impossible because I can't control my feet any more (P8)

Participants indicated that, at a certain point in time, applying adaptation strategies to keep up physical activities was no longer possible. For example, they quit regular group exercises once it became too hard for them to follow the pace of the group: "I gave up Nordic Walking because I could no longer keep up with the group. They always had to wait for me. I'm having to give up more and more things these days. That's a real shame" (P11). When patients were unable to perform certain activities due to a lack of motor skills, they sometimes searched for other, less demanding activities to be able to fulfill a specific social role. For

instance, one participant started volunteering in a nursing home to hand out coffee and tea instead of teaching swimming skills to disabled people. Another started to do maintenance activities at his tennis club instead of being an active tennis player.

Quitting activities also affected professional employment. For many participants, doing their job was too exhausting or the required activities had become physically impossible. They found that sound lower-limb function, an appropriate sitting posture, and sufficient energy level was needed for almost every job, which often made finding an alternative position more difficult.

"I feel ashamed"

Many participants expressed feelings and thoughts about themselves and reflected on how others might think about them. Feeling judged and ashamed was a very significant category of emotions. Another category of emotions was feelings of fear and frustration. Each of these two categories is elaborated below and introduced with a representative quote.

At parties I don't drink much wine, otherwise they think I'm drunk (P11)

Participants regularly encountered negative judgements by persons in their social environment, or felt ashamed of how they walked because of looks or comments from others: "People look at me, they think I'm crazy because I walk so strangely" (P8). The feeling of being observed worsened their gait pattern even more: "When I was on holiday in Rome, I felt much more relaxed walking through the city because nobody knows me there" (P11). Several participants also had to endure other people's opinions, particularly concerning questions about the need for walking aids or mobility devices: "Everyone asks, 'aren't you too young to be using an electric scooter'?" (P2). Among friends, participants could talk freely about spasticity and its consequences in daily life, although they showed less openness amongst others.

I am so scared that I will deteriorate further (P13)

Living with chronic spasticity led to feelings of fear, frustration and depression. Participants feared the slow but inevitable progression of their disease. They tried to stay as active as possible to prevent functional decline. Some avoided the use of walking aids, such as ankle-foot orthoses, because of fears that using such aids might make their muscles even weaker and lead to a more rapid functional decline. Participants experienced feelings of frustration since their social roles gradually became more restricted. Being more homebound also led to having fewer topics to discuss with other people. Some participants found it very difficult to accept the consequences of spasticity, or became angry with themselves when, for example, they could not get out of bed easily. Some even felt depressed, wondering why they were affected by chronic and progressive spasticity.

"I need support"

All participants needed some type of support for dealing with chronic spasticity and its functional consequences. The question on their needs and expectations regarding spasticity management elicited explicit ideas about how participants wanted to be supported by professional healthcare providers. We identified four categories: adequacy of information, efficacy of interventions, quality of healthcare professionals, and insight into disease progression and day-to-day fluctuations. Each category is addressed below and introduced with a representative quote.

I want more information than just the explanation about what the abbreviation HSP means (P5)

Both participants and caregivers mentioned the need of appropriate information about spasticity and its individual consequences: "My partner and I both suffer from the effects of spasticity. We both want to learn more about this" (P5). Participants not only expressed their need of adequate medical information, but also of practical information about how to deal with the consequences of spasticity in daily life. The need for appropriate information was underlined by one patient who stated that she did not want botulinum toxin injections yet, because she was afraid that the effectiveness might wear off over time. Then, when she would really need the injections, they might no longer be effective. Furthermore, she brought up the issue of high costs and the relatively short duration of effects, from which she concluded it would be best to postpone the injections: "Maybe I'll get more out of those injections when I get older. Is it still too soon? … … And even though I don't have to pay for them myself, the prices are shocking" (P13).

Thanks to the injections, the tips of my shoes don't need to be repaired so often (P7)

Regarding the efficacy of interventions, beneficial experiences with botulinum toxin injection were mentioned. Participants experienced a slower progression of spasticity-related problems. A negative effect mentioned by several participants was the initial weakness in their legs after the injections, leading to a wobbling gait and loss of stance stability: "The first few days, my legs are weaker, making me wobblier. I have less control over my legs then" (P10). Participants experienced the benefits of the injections only after the first few days. The effect of botulinum toxin wore off after a time. Participants told that the resulting increase in spasticity-related problems was unpleasant and that they wanted to avoid this. Therefore, some participants preferred a fixed schedule of injections, as they were afraid of receiving their follow-up treatment too late. Some participants did not receive any injections, but were treated with oral spasmolytics. These patients experienced fatigue, sometimes extreme, which they felt was a side effect of the medication. Someone explained that she fell asleep while having visitors, and this was mentioned as a burden.

Many participants performed stretching exercises, either by themselves or supported by a physiotherapist, or a combination of both. Some participants mentioned that they had to stretch their leg muscles daily, since physiotherapy twice a week was not sufficient to reduce their discomfort. Some participants preferred to incorporate stretching exercises into their daily routines: "I stretch my calves at the kitchen worktop while I make my sandwiches for breakfast. That way I won't forget to do those exercises" (P4). Others, however, found that physiotherapists were able to stretch their muscles more effectively than they were able to do themselves. Many considered a combination of physiotherapy and a home-based exercise program to be an optimal solution, as they needed coaching and motivational support to perform the home exercises. Only a few participants did not experience a beneficial influence of physiotherapy in addition to self-treatment. Some experienced no difference when they interrupted their (self-)treatment during the holiday season.

My GP … does not know much about this (P1)

Participants were usually satisfied about the healthcare providers in centers of expertise for HSP, because their treatment was based on a thorough and personalized assessment. Participants felt they were taken seriously, were able to contact the professionals easily,

Table 2. Key messages from the participants.

Theme	Category	Important message
I stumble	Pain and stiffness	Pain and stiffness are cardinal features of HSP.
	Fatigue	Fatigue is a major consequence of spasticity, impacting on both daily routines and night rest.
	Impaired balance and gait	Limitations in gait and balance have an impact on almost all physical activities.
	Day-to-day fluctuations	Within the gradual progression of spasticity, participants experience substantial day-to-day fluctuations.
I struggle	Adjustment to limitations	Spasticity is energy demanding.
	Quitting activities	Giving up activities is common in various domains of daily life. Finding alternatives is a struggle.
I feel ashamed	Feeling ashamed and being judged	Moving differently than others causes feelings of shame and being judged.
	Feelings of fear and frustration	Fear of functional deterioration and frustration about gradually increasing social isolation are common.
I need support	Adequacy of information	There is a need for reliable medical information as well as practical information on how to deal with spasticity.
	Efficacy of interventions	Botulinum toxin injections induce fluctuations of spasticity, which are inconvenient. Physiotherapy can provide coaching and motivation to continue home exercises over time.
	Quality of healthcare professionals	Healthcare providers in expertise centers should support healthcare providers in the community with specific knowledge of how to treat and coach patients with HSP.
	Insight in progression and fluctuations	Monitoring of physical impairments and activity limitations is important to get a better grip on the consequences of lower-limb spasticity on daily life.

and experienced a relationship of trust as most of the professionals reacted promptly and adequately to their questions and needs. In contrast, they were less satisfied with the healthcare providers in the community, both general practitioners and physiotherapists. They felt that most of these professionals were unfamiliar with HSP. They also commented on the absence of a thorough assessment, personalized instruction and treatment, easy access, regular checkups, and shared decision-making. Although treatment close to their homes was convenient, some participants experienced discontinuity or felt that therapists went too easy on them during therapy: "I would really like to receive proper treatment, and not just do the same exercises that I already do at home" (P2).

The symptoms creep up on me, but with a video I could see the difference (P8)

Although participants did not want to be confronted too much with the progression of their disease, they did want to get better grip on their condition by receiving feedback about spasticity fluctuations and the influence of interventions on their possibly fluctuating spasticity. They explained that it would be easier to adhere to the exercise plan if they could notice the effects of their efforts. Furthermore, they thought that objective feedback would provide insight into whether an experienced change was real or not. Monitoring their functional status regarding the joint range of motion, muscle length, gait velocity, gait pattern, pattern of rising and sitting down, and sleep patterns were mentioned as important: "I would like to keep track of the of quality of movement, and make connections between the walking speed, stiffness, and the length of my calf muscles" (P6). Participants felt that the use of wearable devices or short video assessments could be helpful in this respect, yet that monitoring should be undemanding and not be time-consuming. Some participants took their own initiative to monitor their experiences after change of medication: "When I switched from baclofen to botulinum toxin, I made a note of the changes I noticed in my phone" (P4).

Discussion

This qualitative study investigated the experiences and needs of patients with chronic spasticity due to a pure form of HSP at the level of physical activities, social participation, and general well-being. Based on semi-structured interviews with 14 purposively selected patients, four common themes were identified that are reflected by the following phrases: (1) "I stumble", (2) "I struggle", (3) "I feel ashamed", and (4) "I need support". Participants literally or metaphorically stumbled when performing a wide range of physical activities. Applying adequate strategies to manage spasticity and its consequences was often perceived as a struggle in terms of the medical, role, and emotional management [16]). Furthermore, participants often felt ashamed as they experienced negative judgements by persons in their environment. Finally, they expressed the need for medical and practical support not only from professionals working in centers of expertise, but also from community healthcare professionals. In Table 2, we have summarized some of the important messages from the participants.

Spasticity is more than just stiffness

Participants reported some well-known physical consequences of chronic lower-limb spasticity, such as stiffness, leg pain, and a stumbling gait pattern [9,17]. In addition, they mentioned less obvious consequences of lower-limb spasticity, such as back pain, fatigue, and unpredictable day-to-day fluctuations. Besides leg pain, nearly all participants complained about lower back pain, which they believed to be due to an altered posture and gait pattern. They also experienced that both a strenuous gait and a disturbed sleeping pattern were responsible for high levels of fatigue. Together, these symptoms had a huge impact on their daily lives. Notably, pain and fatigue have been scarcely addressed in the literature on HSP, even though they are regarded by the patients as some of their most disabling symptoms [18]. This observation underscores the importance of monitoring fatigue and pain levels in persons with chronic spasticity, and of integrating energy and pain management strategies into the rehabilitation regimens for people with HSP.

The need to address outcomes that are meaningful to the patient

Cusick et al. [7] argued that the most frequently used measurement tools in the rehabilitation of chronic spasticity are focused on the level of physical impairments, particularly measures of muscle tone and joint range of joint motion [7]. Although these are relevant outcome measures from a medical perspective, they do not address the issues that are most relevant to the patients, such as fatigue, pain, sleep, and general well-being. Outcomes that address fatigue, energy expenditure, and sleep appear to be lacking in the literature on chronic spasticity in persons with HSP. Incorporating such measures into the treatment regimens for

chronic spasticity may help to tailor interventions to the needs of individual patients.

In addition to physical impairments, all participants experienced serious limitations across a wide range of physical activities, including the domains of personal care, domestic life, employment, and leisure. Apart from the publication by Grose et al. [17], who found similar limitations, the literature on HSP has scarcely addressed the extent to which chronic spasticity impacts on these physical activities, and how patients contend with their physical activity limitations and related participation restrictions. According to the new definition of health by Huber et al. [19], health is not the absence of disease, but the ability to adapt to the consequences of disease and being able to self-manage these consequences [19]. This definition implies that healthcare providers should not only assess physical impairments and the manner in which these are managed by patients (medical management), but should also systematically evaluate the performance of daily activities and the level of participation to understand the impact of spasticity on the daily life of the individual patient with HSP (role and emotional management).

Against the above-mentioned background, measuring relevant outcomes identified by persons with chronic spasticity, referred to as patient-reported outcomes (PROs), can be helpful to tailor interventions to individual needs [20] and to develop personalized monitoring strategies throughout the course of the disease. At present, patient-reported outcome measures (PROMs) that have been developed for and used by people with chronic hereditary spasticity are lacking, although the Spastic Paraplegia Rating Scale (SPRS), validated for patients with hereditary spastic paraplegia [21], may be a valuable alternative tool. This study has identified several functional aspects, such as spasticity-related fatigue, pain, sleeping problems, and activity limitations, for which PROMs must be developed.

Personal feedback enhances individual grip on spasticity

Participants explained that they had difficulties gaining control of the changes in their spasticity levels, either changes that occur spontaneously or those induced by interventions. For instance, participants found it difficult to differentiate between disease progression and the wearing-off phenomenon following focal spasmolysis. In addition, participants reported that both internal factors (stress, fatigue) and external factors (temperature) could influence spasticity. To enhance the patients' control over spasticity, they expressed the need to have insight into the relevant changes that occur over time. Systematic monitoring of PROs may help both patients and professionals to monitor disease progression and to evaluate the effect of interventions [20]. In addition, professionals should meticulously monitor the status of several bodily functions and capacities, because slight but gradual changes in, for example, muscle length may eventually have a negative impact on the performance of activities.

The monitoring of PROs would seem to be a mutual responsibility between the professionals and patients. Indeed, the benefits of self-monitoring have already been demonstrated: patients with chronic obstructive pulmonary disease and in patients with cerebral palsy were able to detect relevant changes in their health condition [22], which reduced long-term complications [23].

Empowering patients' capacity for adaptation and self-management

Our participants reported a difference between healthcare professionals in expertise centers and those in community-based centers. Particularly, they missed sufficient expertise when they consulted their general practitioner and/or physiotherapist in the community. They felt that treatment quality could be enhanced by more knowledge of the consequences of spasticity in everyday life. In addition, they needed more personalized instruction and coaching on how to adapt to these consequences (role and emotional management). Therefore, it seems important that healthcare providers in expertise centers exchange their knowledge with healthcare providers in the community to avoid a mismatch regarding goals and priorities for rehabilitation.

Such mismatches have been reported for the stroke population [24] and for persons with Parkinson's disease [25]. For example, in a systematic review, Jellema et al. [26] showed that public belief and support from others were crucial for patients with stroke to overcome their barriers to perform certain activities. In addition to the education of stroke patients regarding home, work, and community activities and possible adaptation strategies, they recommended providing stroke survivor's proxies with the knowledge and skills to encourage patients to resume their social participation [26]. From this perspective, patients with HSP and their close kin should also be taught adaptation strategies to reduce the consequences of chronic spasticity in their daily lives. To achieve this aim, an interdisciplinary approach may be necessary including contributions from occupational therapists and psychosocial disciplines.

Strengths and limitations

This study has a number of strengths. We used the COREQ criteria for reporting qualitative research [14]. The data were collected by means of interviews at the participants' homes, which created a comfortable and safe atmosphere. A member check was performed by presenting the conversational notes to the participants and allowing them to check the accuracy. The procedure for data collection and analysis as well as the results were reviewed together with the second author, and eventually with the entire research team. A limitation of this study is that intimate topics, such as bladder issues or sexuality, although mentioned in the literature [9], were not addressed by our participants. Reasons for not mentioning these topics by our participants might be feelings of shame and the possibility that they associated spasticity primarily with impairments of the lower limbs rather than with bladder or sexual dysfunction. Another limitation is that the results remain limited to patients with a pure form of HSP, and do not include the more complicated subtypes. As a consequence, the results cannot be generalized to other HSP populations.

Conclusion

This study shows that the consequences of chronic lower-limb spasticity in patients with pure forms of HSP go beyond the well-known consequences such as muscle stiffness and impaired ambulation. HSP is associated with fatigue, pain, unpredictable day-to-day fluctuations, a wide range of physical activity limitations and social participation restrictions, and the continuous need to adjust compensation strategies to overcome these limitations. In addition, feelings of shame, fear, frustration, and depressed mood coincide with the disabilities experienced and the judgements made by persons in the social environment. The participants expressed a strong need for interventions not only to reduce the neuromuscular impairments, but also to alleviate fatigue and pain, to improve nighttime rest, and to restore the capacity to perform relevant physical activities and fulfill social

roles. Spasticity rehabilitation regimens should therefore be tailored to the individual needs of the person with HSP, based on careful personalized monitoring that includes patient-reported outcomes. Enhancing self-management may empower patients to gain control of the consequences of chronic spasticity in their daily lives.

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ORCID

Hans C. J. W. Kerstens http://orcid.org/0000-0002-6492-4590
Bas J. H. Van Lith http://orcid.org/0000-0001-6609-8782
Alexander C. H. Geurts http://orcid.org/0000-0001-7478-1119
Maria W. G. Nijhuis-van der Sanden http://orcid.org/0000-0003-2637-6877

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